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Editorial Comment

Pulmonary arteriovenous fistulas in patients with left isomerism and cardiac malformations

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OVER THE PAST DECADE, VARIOUS FORMS OF cavopulmonary anastomoses (the bidirectional Glenn shunt, the hemi-Fontan procedure) have become the palliative procedure of choice for the treatment of hearts with a functionally single ventricle. Significantly, the occurrence of pulmonary arteriovenous fistulas had been noted as an important 'late' complication after construction of the classic Glenn anastomosis, with a reported incidence of 25%.[1] In contrast to the experience with the classic anastomosis, the incidence of fistulas after construction of bidirectional cavopulmonary anastomoses seems to be quite low. These observations, however, may simply be the consequence of the short natural history of the bidirectional shunt. In most instances, it is performed as a part of a two-stage approach, with most patients proceeding relatively quickly to the Fontan operation, which then incorporates the inferior caval and hepatic venous flows into the pulmonary circulation. An exception to this two-stage strategy for total cavopulmonary connection is in patients with left isomerism, interruption of the inferior caval vein, and azygos continuation to the superior caval vein, in which the hepatic veins drain directly into the atrium. In these cases, construction of a bidirectional cavopulmonary anastomosis directs all venous return to the pulmonary bed except for the coronary and the hepatic venous blood. This, typically, results in arterial oxygen desaturation in the range of 85% to 90%, and was initially considered to provide definitive palliation.

Several years ago, however, some cases of fistulous communications were reported in patients with left isomerism who had not undergone previous surgery, suggesting that the isomerism in itself may predispose to the development of the pulmonary arteriovenous connections.[2,3] A further study[4] then showed a high incidence of fistulas (21%) in patients with left isomerism after a bidirectional cavopulmonary anastomosis had been constructed as definitive palliation. The abnormal distribution of pulmolnary blood flow to the upper and lower lobes, and the lack of pulsatile flow, were initially evoked as causative factors. This hypothesis, however, is unsatisfactory, because similar flow dynamics exist after the Fontan operation, where it is unusual to find pulmonary arteriovenous fistulas.

In most of the patients reported in the literature, in fact, the common anatomic feature is the exclusion of normal hepatic venous flow from the affected lung. The hypothesis that the diversion of normal hepatic venous flow from the pulmonary circulation plays a role in development of fistulas is also supported by the findings of similar changes in adults with cirrhosis, and in children with biliary atresia. Furthermore, the lesions may then regress after liver transplantation.[5] This suggested the existence of a biochemical hepatic product resulting from normal hepatic metabolism of a stimulatory factor, or synthesis of a suppressor factor, as a cause of formation of the fistulas. This seemed to be confirmed by the reversibility of the change after rerouting the hepatic blood flow to the pulmonary beds.[6]

On the other hand, the evidence for development of fistulas in patients with left isomerism without cardiac anomalies, the strong relationship between the biliary atresia and fistulas,[7] and some reports of aggravation of fistulas after conversion of a bidirectional cavopulmonary shunt to a total cavopulmonary anastomosis,[8] suggest that the hepatic venous blood is playing some part, but not

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the exclusive role in the genesis of fistulous formation. The finding of systemic arteriovenous fistulas,[9] and systemic veno-venous malformations (porto-caval collaterals, intrahepatic malformations) in patients with left isomerism suggests also that the postulated vasoactive agents are affecting other organs in association with the lungs. It is possible to hypothesize that, in these patients, vasodilator agents (such as glucagon, substance P, vasoactive intestinal peptide, and so on), which originate from the mesenteric circulation, fail to be metabolized by the liver due to the porto-caval veno-venous collaterals. Instead, they pass directly to the lungs, the heart and the other organs, then stimulating the formation of arteriovenous fistulas. An overly-sensitized pulmonary endothelium to an hepatic or mesenteric factor, or a reduced sensitivity of the vasculature to endogenous vasoconstrictor, may also be considered as alternative mechanisms.

Thus, while a non-random association is unequivocally established between left isomerism and pulmonary arteriovenous fistulas, the exact pathogenetic mechanism for formation of the fistulas is still not completely understood.

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